Papuloerythroderma of Ofuji – A Case report

Dr. Ashutosh Chate[1], Dr. Vishal Indurkar[2], Dr. Ramesh Gosavi[3], Dr. Nausheen Syed[4], Dr. Aishwarya Kitture[5]

[1,4,5]Resident, [2]Associate Professor, [3]Professor & Head, Department of Dermatology, DVVPFs Medical College & Hospital, Ahmednagar-414111

Corresponding Author : Dr. Ashutosh Chate

E-mail : ashutoshchate@gmail.com

Address : Department of Dermatology, DVVPFs Medical College & Hospital, Ahmednagar-414111

Abstract :
Papuloerythroderma of Ofuji is disease of unknown etiopathogenesis It is clinically characterised by generalised intensely pruritic eruption of flat topped red to brown coalescing papules with sparing of skin folds and creases. It usually presents in elderly and is associated with malignancies, infection and drugs. Here we report a case of Papuloerythroderma of Ofuji with characteristic clinical features of idiopathic type.

Key words : Papuloerythroderma, Eosinophilic dermatosis.

Introduction:

Papuloerythroderma of Ofuji is a rare disease was first described by Ofuji et al.[(1)] Based on their observations, the authors made an etiological division of the disease into 4 forms:

1. Primary (idiopathic)
2. Secondary (in the course of atopic disease, neoplasms, infections, drugs)
3. Papuloerythroderma imitating lymphoma
4. Other diseases with a clinical picture similar to that of erythroderma (erythroderma without papules)

It usually affect elderly men and is characterized by generalized and intensely pruritic eruption of flat-topped red-to-brown coalescing papules with sparing of skin folds and creases.[(2)] Etiopathogenesis remains enigmatic but proposed pathogenesis involves Th2 lymphocytes directed toward specific/ unknown antigen, which can be neoplastic, microbial, pharmacological, or self-antigen. These lymphocytes produce soluble mediators which induce the proliferation, activation, and migration of eosinophils and other cell types into the skin, which then enhance the cutaneous inflammation.[(3)]

Case Report: A 55-year-old male presented with black colored raised lesions over whole body since more than 3 years. The lesions were associated with intense itching. Patient treated symptomatically but the lesions never subsided completely. There was no history of contact with parthenium (congress grass in local language), no history of chronic sun exposure, no history of chronic drug intake, no history of personal/ family history of atopy, no history of weight loss, no history of medical illness and similar complaints in the family. His general physical & systemic examination was unremarkable. There was no significant lymphadenopathy. On cutaneous examination, there were multiple red brown, flat-topped papules coalesces to cover entire body. Lesions were predominantly distributed over extensors with sparing of skin folds and flexures called as deck-chair sign.**(Figure 1 & 2)** Oral mucosa and hair were normal and nails were shiny in appearance. On history & clinical examination we kept the possibility of phytophotodermatitis, Atopic dermatitis and Mycosis fungoides. Patient was investigated thoroughly with Complete hemogram revealed leucocytosis and eosinophilia with (25% eosinophils). His liver function test, renal function test, random blood sugar were normal and chest radiographs, ultrasound of abdomen/ pelvis also were within normal limits. Skin biopsy taken for histopathological examination shows moderately dense superficial perivascular lymphocytic infiltrate of lymphocytes and many eosinophils with spongiotic psoriasiform changes in the overlying epidermis and absence of atypical lymphocyte and epidermotrophism. **(Figure 3)** Histopathological findings were suggestive of papuloerythroderma of Ofuji and Phytophotodermatitis. On correlation of clinical feature and histopathological findings diagnosis of papuloerythroderma of Ofuji was made. Patient was started on oral prednisolone 40 mg daily, which was gradually tapered, along with azathioprine 50 mg, twice daily, Antihistamine, and topical corticosteroid. There was significant improvement at the end of 4 weeks, and patient is on further follow up.**(Figure 4)**

Figure 1 & 2: Red brown colored coalescing papules covered trunk & extremities along with sparing of skin folds & flexures i.e. Deck chair sign positive

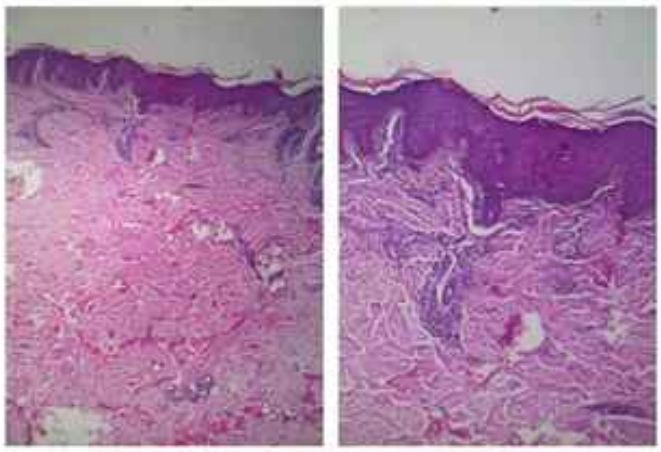

Figure 3: moderately dense superficial perivascular lymphocytic infiltrate of lymphocytes and many eosinophils and absence of atypical lymphocyte and epidermotrophism.

Discussion:

Papuloerythroderma of Ofuji clinically presents with multiple polygonal erythematous papules that spare the body folds. It is accompanied with intense pruritus, lymphadenopathy, peripheral eosinophilia, and raised serum IgE level.[2] Deck-chair sign, that is, sparing of abdominal folds, is highly characteristic of papuloerythroderma of Ofuji but has also been observed in erythroderma owing to atopic dermatitis, psoriasis, imatinib-induced rash, cutaneous Waldenstrom's macroglobulinemia, angioimmunoblastic T-cell lymphoma, acanthosis nigricans, erysipelas, discoid lupus erythmatosus, contact dermatitis, and leprosy, and can be idiopathic.[3-5] Histopathology consists of mild hyperkeratosis, acanthosis, focal spongiosis, and perivascular lymphocytic infiltration, with eosinophils in the upper dermis with absence of atypical lymphocytes.[6]

Torchia and colleagues proposed the diagnostic criteria of the disease.**(Table 1)** In our patient, all the major and minor criteria were fulfilled except peripheral lymphopenia. There was no underlying predisposing factor in our patient. Diagnosis of idiopathic form of the syndrome requires the presence of all the 5 major criteria. Similar cases from India have been reported by Prasad et al[2] and Gulati.[7] Treatment modalities include psoralen ultraviolet-A therapy, topical and systemic steroids, etretinate, cyclosporine, and interferon[8], we have started patient on azathioprine 50mg twice a day with noticeable improvement in 4 weeks. **(Figure 4)**

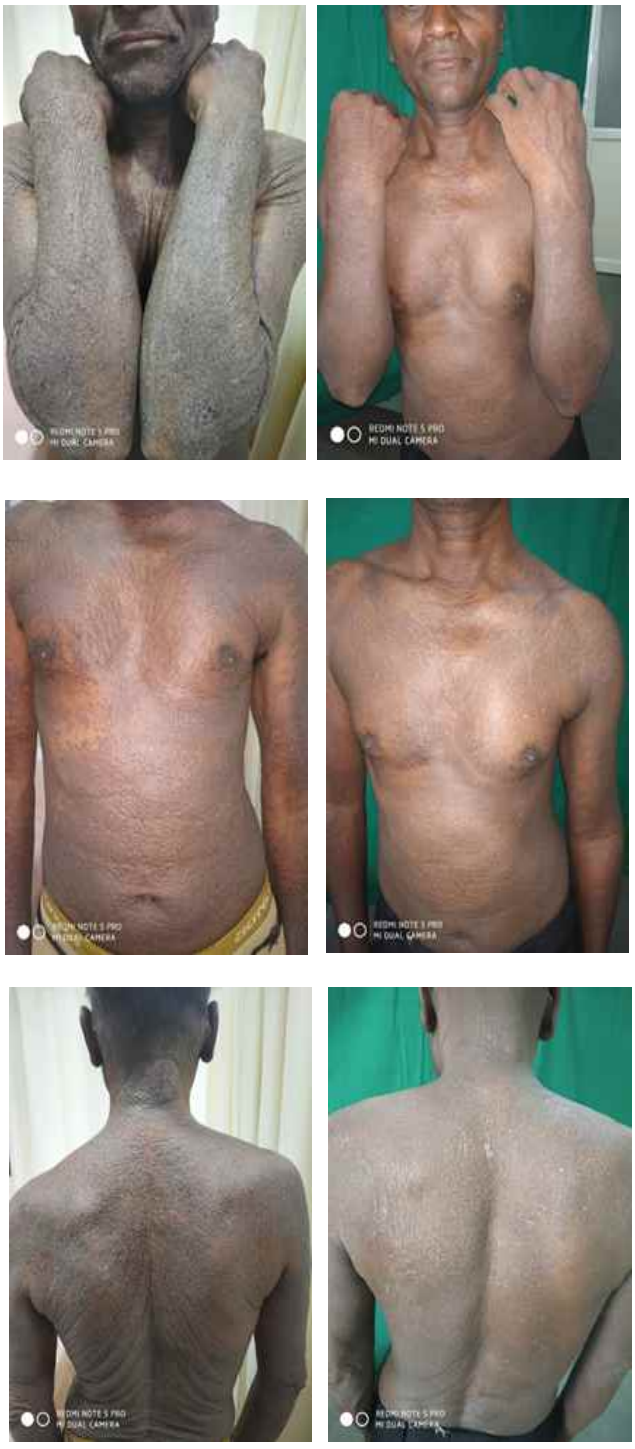

Figure 4: Pre & Post treatment with Azathioprine therapy

Table 1: Torchia et al Diagnostic Criteris of Papuloerythroderma of Ofuji[3]

Major Criteria	Minor Criteria
1. Erythrodermic eruption, consisting of flat -topped, coalescent, red-brown papules with a cobblestone-like appearance 2. Pruritus 3. Sparing of skin folds creases 4. Histopathological exclusion of cutaneous lymphoma & other skin diseases 5. Absence of the causative factors such as tumors, infections, drugs & atopy	1. Age greater than 55 years 2. Male sex 3. Peripheral and /or tissue eosinophilia 4. Elevated level of total IgE 5. Peripheral lymphopenia

Conclusion:

As this disease is associated with internal malignancy and cutaneous T-cell lymphoma, proper follow-up of these patients should be done even after remission of cutaneous lesions. In medical practice we should always ready for surprises.

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